12/3, K, AB/2 (Item 2 from file: 155) DIALOG(R) File 155: MEDLINE(R)

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05289096 PMID: 3738904 86290372

Citrullinemia: quantitative deficiency of argininosuccinate synthetase in the liver.

Oyanagi K; Itakura Y; Tsuchiyama A; Nakao T; Nakano K; Saeki T

Tohoku journal of experimental medicine (JAPAN) Apr 1986, 148

(4) p385-91, ISSN 0040-8727 Journal Code: 0417355

Document type: Journal Article

Languages: ENGLISH

Main Citation Owner: NLM Record type: Completed

Two cases of citrullinemia were reported. Case 1 was an one month old female. Her clinical course and findings were different from the fulminant neonatal citrullinemia reported in predominantly Caucasian of countries. Our patient was well controlled under a low protein diet and essential amino acids till 9 months of age, but unfortunately she died of Reye's like syndrome. Case 2 was 31 year old male (at the time of death). He was admitted to our hospital because of hyperammonemia and mental retardation. By subsequent laboratory investigations he was diagnosed as adult type of citrullinemia and died of hepatoma . Enzymological analysis revealed that argininosuccinate synthetase (ASS) activities in the liver tissues of the patients decreased to 40% (Case 1), 20% (Case 2) compared with those in control liver tissues. The other urea cycle enzyme activities were all within normal range. ASS activities in the kidney and brains of the two cases were within normal range. The kinetic constant values of ASS for three substrates in the tissues of liver and kidney were all normal. Results of immunochemical analyses indicated that citrullinemia in our patients was caused by a quantitative deficiency of ASS associated proteins of the liver and kidney tissues as to the molecular weight.

16/20

1990, Chemical

Name:

Tumor

Markers,

Biological; Urea; Arginine;

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Set
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      2706475
S2
           64
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S3
            6
                S1 AND S2
S4
                RD (unique items)
                (LACK? OR REDUC? OR DEFICIENT) (5N) ARGININE
S5
         4986
S6
          306
                S1 AND S5
S7
          267
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S8
          174
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S9
          166
                S8 AND PY<2001
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         1203188 CANCER
         1307639
                  TUMOR
          738210 CARCINOMA
          132182 MELANOMA
           40858 HEPATOMA
            2189 ARGININOSUCCINATE
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                  (5N) ARGININOSUCCINATE
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Processing
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                 PY<=2001
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              46 S11 AND PY<=2001
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DIALOG(R) File 155: MEDLINE(R)
(c) format only 2003 The Dialog Corp. All rts. reserv.
06840618
           91080593
                      PMID: 2259230
  Elevated argininosuccinate synthetase activity in adult T leukemia cell
lines.
  Sugimura K; Kimura T; Arakawa H; Ohno T; Wada Y; Kimura Y; Saheki T;
Azuma I
  Institute of Immunological Science, Hokkaido University, Sapporo, Japan.
             research (ENGLAND) 1990, 14
  Leukemia
                                                 (10)
                                                         p931-4, ISSN
           Journal Code: 7706787
0145-2126
  Document type: Journal Article
  Languages: ENGLISH
  Main Citation Owner: NLM
  Record type: Completed
  Argininosuccinate synthetase (ASS) is a ATP-dependent and rate-limiting
enzyme of the urea cycle which catalyzes L-citrulline to L-arginine in
combination with argininosuccinate lyase (ASL). We demonstrate here that
(a) human normal T and B lymphocytes did not express ASS activity, (b)
however, three adult T leukemia (ATL) cell lines tested here exhibited
significant elevation of ASS activity, and (c) ASL activity remained
relatively constant in normal lymphocytes and various leukemia cell lines.
These results suggest that the ASS expression of peripheral blood lymphocytes is of value as a diagnostic marker of leukemia including ATL.
The implication of these results is discussed.
```

Argininosuccinate Lyase; Argininosuccinate Synthase

12/3,K,AB/2 (Item 2 from file: 155)

DIALOG(R) File 155: MEDLINE(R)

(c) format only 2003 The Dialog Corp. All rts. reserv.

05289096 86290372 PMID: 3738904

Citrullinemia: quantitative deficiency of argininosuccinate synthetase in the liver.

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Tohoku journal of experimental medicine (JAPAN) Apr 1986, 148

(4) p385-91, ISSN 0040-8727 Journal Code: 0417355

Document type: Journal Article

Languages: ENGLISH

Main Citation Owner: NLM Record type: Completed

Two cases of citrullinemia were reported. Case 1 was an one month old female. Her clinical course and findings were different from the fulminant neonatal citrullinemia reported in predominantly Caucasian countries. Our patient was well controlled under a low protein diet and essential amino acids till 9 months of age, but unfortunately she died of Reye's like syndrome. Case 2 was 31 year old male (at the time of death). He was admitted to our hospital because of hyperammonemia and mental retardation. By subsequent laboratory investigations he was diagnosed as having adult type of citrullinemia and died of hepatoma . Enzymological analysis revealed that argininosuccinate synthetase (ASS) activities in the liver tissues of the patients decreased to 40% (Case 1), 20% (Case 2) compared with those in control liver tissues. The other urea cycle enzyme activities were all within normal range. ASS activities in the kidney and brains of the two cases were within normal range. The kinetic constant values of ASS for three substrates in the tissues of liver and kidney were all normal. Results of immunochemical analyses indicated that citrullinemia in our patients was caused by a quantitative deficiency of ASS associated proteins of the liver and kidney tissues as to the molecular weight.

Apr 1986,

... subsequent laboratory investigations he was diagnosed as having adult type of citrullinemia and died of hepatoma. Enzymological analysis revealed that argininosuccinate synthetase (ASS) activities in the liver tissues of the patients decreased to 40% (Case 1...

12/3,K,AB/3 (Item 3 from file: 155) DIALOG(R)File 155:MEDLINE(R)

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? s cancer or carcinoma or tumor or melanoma or hepatoma or malignan?
Processing
         1203188 CANCER
          738210 CARCINOMA
         1307639 TUMOR
          132182 MELANOMA
           40858 HEPATOMA
          493621 MALIGNAN?
      S1 2706475 CANCER OR CARCINOMA OR TUMOR OR MELANOMA OR HEPATOMA OR
                  MALIGNAN?
? s (reduc? or lack? or deficient) (5n) (argininosuccinate)
         3293688 REDUC?
          548491 LACK?
          232061 DEFICIENT
            2189 ARGININOSUCCINATE
              64 (REDUC? OR LACK? OR DEFICIENT) (5N) (ARGININOSUCCINATE)
      S2
? s s1 and s2
         2706475 S1
              64 S2
      S3
               6 S1 AND S2
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DIALOG(R) File 155: MEDLINE(R)
(c) format only 2003 The Dialog Corp. All rts. reserv.
07626920
          93082106
                     PMID: 1450673
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5/3,K,AB/2 (Item 2 from file: 155)
DIALOG(R)File 155:MEDLINE(R)
(c) format only 2003 The Dialog Corp. All rts. reserv.

09326325 21083439 PMID: 11215740

. Hepatocellular carcinoma associated with adult-type

citrullinemia.

Ito T; Shiraki K; Sekoguchi K; Yamanaka T; Sugimoto K; Takase K; Tameda Y

First Department of Internal Medicine, Mie University School of Medicine, Tsu, Japan.

Digestive diseases and sciences (United States) Nov 2000, 45

(11) p2203-6, ISSN 0163-2116 Journal Code: 7902782

Document type: Journal Article

Languages: ENGLISH

Main Citation Owner: NLM Record type: Completed

Hepatocellular carcinoma associated with adult-type

citrullinemia.

Nov 2000,

Descriptors: Carcinoma, Hepatocellular--pathology--PA

6/hd

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? s citrullinemia
     S1 584 CITRULLINEMIA
? s cancer or tumor or carcinoma or melanoma or hepatoma
        1203188 CANCER
        1307639 TUMOR
         738210 CARCINOMA
         132182 MELANOMA
          40858 HEPATOMA
     S2 2510740 CANCER OR TUMOR OR CARCINOMA OR MELANOMA OR HEPATOMA
? s s1 and s2
        584 S1
2510740 S2
             16 S1 AND S2
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>>>Duplicate detection is not supported for File 340. .
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...completed examining records
          12 RD (unique items)
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Processing
Processing
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       40178892 PY<=2001
      S5 10 S4 AND PY<=2001
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 5/3,K,AB/1
DIALOG(R) File 155: MEDLINE(R)
(c) format only 2003 The Dialog Corp. All rts. reserv.
                    PMID: 11
09796859
           21604463
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